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Case report

Recurrent disseminated encephalomyelitis: A case report and literature review

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ABSTRACT

Background: Acute disseminated encephalomyelitis has been understood as a monophasic, often post-infectious illness that predominantly affects the pediatric population. Though that describes the majority of cases, exceptions do exist. In this case report, we present an adult case of recurrent disseminated encephalomyelitis (DEM) and review the available literature on this clinical entity.

Methods: PubMed search performed using the terms "MDEM" and "Recurrent ADEM" in April 2018. A total of 23 items resulted for the first search and another 142 for the second. We selected articles that described cases of recurrent ADEM with a preference for those publications describing adult cases and those written in English language.

Conclusion: Recurrent disseminated encephalomyelitis is a distinct clinical entity that has features which overlap with multiple sclerosis, making it imperative to distinguish the two. Our case presentation and accompanying literature review highlights the limited scope of data available on recurrent DEM and the need for further study.

1. Introduction

Historically, acute disseminated encephalomyelitis has been understood as a monophasic, often post-infectious illness that predominantly affects the pediatric population. Though that describes the majority of cases, exceptions do exist. In this case report, we present an adult case of recurrent disseminated encephalomyelitis (DEM). We also performed a literature review in PubMed using the terms “MDEM” and “Recurrent ADEM” in April 2018. A total of 23 items resulted for the first search and another 142 for the second; the timing of these publications spanned from October 1996 to November 2017 and July 1971 to February 2018, respectively. We selected articles that describe cases of recurrent ADEM with a preference for those publications describing adult cases and those written in English language.

2. Case report

A 51-year-old woman was brought to the emergency department by her family for evaluation of bizarre behavior over the preceding two days. She carried diagnoses of diabetes and hypertension and her family history was notable for multiple sclerosis in her mother. Her family denied a history of alcohol or illicit drug use. She had developed symptoms of an upper respiratory infection and had become less interactive over several days. Her exam was notable for orientation to person only and an inability to follow commands. She was spontaneously moving all four extremities and tracking her eyes in all directions. Reflexes were 2 + and symmetric with flexor plantar response. Initial CT brain showed patchy parenchymal hypodensities in periventricular and subcortical distribution concerning for demyelination. Subsequent MRI brain (Fig. 1) revealed numerous enhancing lesions throughout bilateral supratentorial periventricular and subcortical white matter. Cerebrospinal fluid (CSF) and serologic testing results are in Table 1. CT chest, abdomen and pelvis obtained to evaluate for malignancy showed 6 cm cystic lesion in right adnexa and less than 2 mm pulmonary nodule in left lung. The adnexal mass was biopsied and found to be benign. She was diagnosed with acute disseminated encephalomyelitis (ADEM) and initially treated with IV methylprednisolone 250 mg every 6 h for 5 days. She improved modestly but was still not at baseline, prompting further treatment with IVIg at 1 g/kg for two days. At a six-month clinic follow up with a neuroimmunologist, she had returned to baseline.

Ten years later, she was again brought to the emergency department for evaluation of five days of gradually progressive left sided weakness, headache, dizziness and vertigo complicated by one fall. She had minimal cognitive or personality changes. MRI brain revealed multiple enhancing bilateral white matter lesions, similar in distribution to prior

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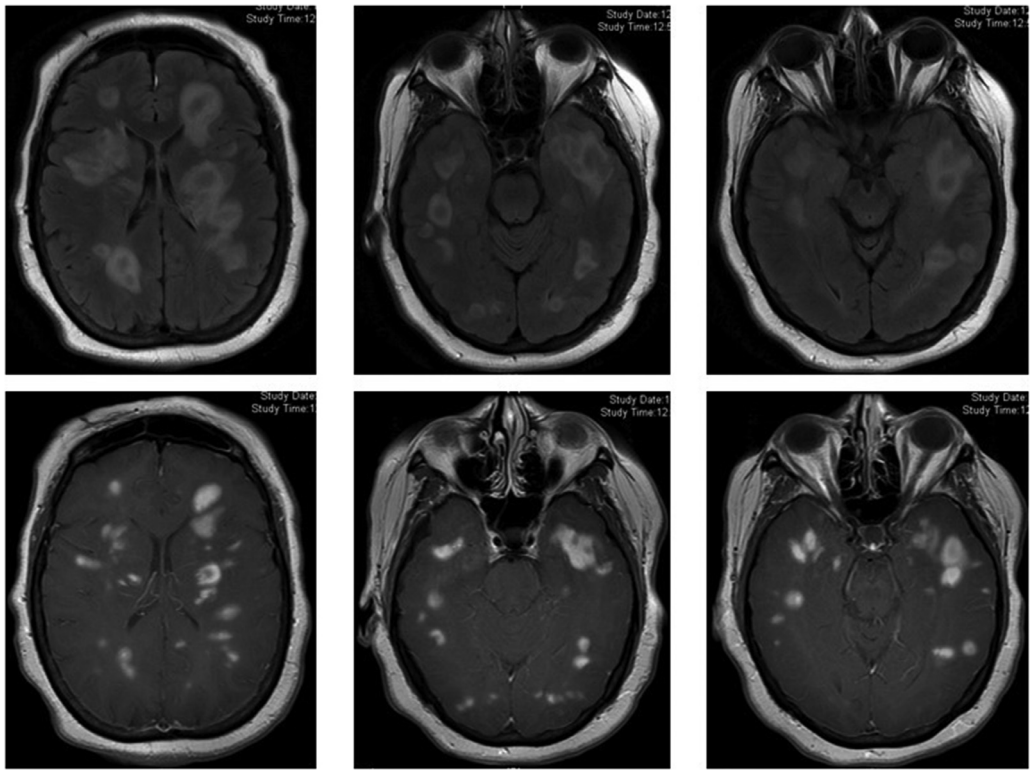

Fig. 1. MRI Brain FLAIR sequences (top row) and images after administration of contrast (bottom row) during initial presentation with encephalopathy. Evidence of numerous enhancing T1 hypointense lesions with surrounding edema in the bilateral supratentorial, periventricular, and subcortical white matter.

Table 1
Serum and cerebrospinal fluid study results.

	Initial presentation, 2008		Recurrent presentation, 2018	
Serum/Urine studies				
Urine toxicology screen	+benzodiazepine, THC, cocaine		Negative	
Serum toxicology screen	Negative		Negative	
Ammonia (16–50 umol/L)	27			
TSH (0.34–5.66 uIU/mL)	1.83		1.39	
HIV	Negative		Negative	
RPR	Non-reactive		Non-reactive	
ANA	1:160- Low titer positive		–	
ANCA	Negative		–	
Anti-SM, RNP, Ro, La	Negative		–	
ESR (0–15 mm/h)	22		7	
C- reactive protein (< = 0.60 mg/dL)	–		0.47	
Serum ACE (7–46 U/L)	3		–	
Lyme IgG	Negative		–	
Paraneoplastic Autoantibody Evaluation	Negative		–	
HHV-6 PCR	Negative		–	
JC Virus PCR	–		Negative	
Vitamin B12 (123–730 pg/mL)	–		274	
CSF studies				
	Tube 1	Tube 4	Tube 1	Tube 4
WBC	11	9	0	1
Neutrophils (%)	92	3		
Lymphocytes (%)	5	81		
Monocytes (%)		10		
Macrophages (%)	3	3		
Variant lymphocytes (%)		3		
RBC	29	11	1450	568
Protein (15–50 mg/dL)	63		82	
Glucose (mg/dL)	71		115	
Bacterial culture	Negative		Negative	
Meningitis panel	–		Negative	
Oligoclonal Band, IgG index	1		2	
IgG Index	0.45			
Paraneoplastic panel	No autoantibodies detected			
Cytology/Flow	Negative for malignancy		Negative for malignancy	
ACE (0.0–2.5 U/L)	<4.0		1.0	